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Orbital Compression Syndrome in a Nigerian Child with Sickle Cell Anaemia: Treatment Challenges in a Resource-Poor Setting.

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ABSTRACT

Background: Sickle cell disease is a relatively common genetic disorder in the sub-Saharan Africa. Orbital compression syndrome (OCS) as one of its various complications, is uncommon. It is however potentially serious, as it impairs vision and can lead to death. This report presents the review of a Nigerian child who presented in the emergency unit of a tertiary hospital with vaso-occlusive crises (bone pain) from sickle cell anaemia. The crises evolved to involve the orbits in this uncommonly reported manifestation.

This diagnosis must therefore alwaysbe considered a potential possibility in all centres involved with care of sickle cell children, especially in low resource settings with limited or unavailable hi-tech investigations. It should be tackled promptly and aggressively, to avert blindness or death.

Key words: Sickle cell anaemia, orbit, complication, child

I. INTRODUCTION:

Vaso-occlusive crisis is the most common complication of sickle cell disease. Ophthalmologic complications of sickle cell disease are unusual findings and can affect any part of the eye and surrounding bones with potential loss of vision.[1-4]Orbital Compression Syndrome (OCS) usually manifests following vaso-occlusive crisis in which the orbital bones are infarcted leading to inflammation and orbital swelling.[2-6]It should be suspected in patients with acute painful periorbital swelling. Orbital compression syndrome with possible optic nerve injury is an uncommon but serious complication. Therefore, this diagnosis should be viewed with a high index of suspicion. It has been reported by different authors.[1-6]

Symptoms include facial pains, headache, fever and poor vision.There is usually restriction of eye movement and proptosis.Subperiosteal haematoma is usually seen on Computed Tomographic Scan or Magnetic Resonance Imaging of the skull around the orbit[4-5].Treatment is usually supportive with intravenous fluids, methyl prednisolone, analgesics and antibiotics. Surgical evacuation of large haematomas isneedful in some cases. Timely treatment is crucial for speedy recovery of the patient and prevention of blindness[3-6].

This report aims to highlight the importance of maintaining a high clinical index of suspicionespecially in resource-poor settings, in children with known sickle cell disease presenting with pain, orbital swelling and restriction of ocular movement. Timely red blood cell transfusion only may be sufficient for uneventful recovery, in some cases without steroids or surgical intervention.[3-6]

II. CASE REPORT

A 9-year-old male was brought in, tothe Children Emergency Unit of this tertiary hospital,witha four day history of fever, paleness of the body,and joint pains respectively.Fever was low grade, intermittent and temporally relieved by Paracetamol tablet. Paleness of the body was gradual in onset, noticed as progressive whitening of the palms and soles of the feet. Joint pains were felt on the knees, shoulders, and wrist. There were no known aggravating or relieving factors and no joint swelling. He had been treated for malaria, with an artemisinin combination drug bought over the counter. When symptoms worsened, he was brought to the children emergency unit of the hospital, for further evaluation.

He was first diagnosed of sickle cell anaemia at the age of four years, and had been on several hospitalizations on account of sickle cell crises, mostly vaso-occlusive. He had also received some red cell transfusions, with the last transfusion being two years before the present admission.

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There was no history of any surgery. He was inconsistent at follow up in the Paediatric haematology clinic, therefore his steady state Packed Cell Volume (PCV) was not known. He had been given full childhood immunizations.

He lived with his parents in a monogamous family setting, and was the youngest of four children. He was the only child of the four, who had sickle cell anaemia. His mother was a 45-year-old teacher with tertiary level of education and his father is a 52 year old journalist, also with tertiary level of education.

On general examination at presentation, he was conscious, but lethargic and in painful distress. He had long thin extremities and gnathopathy, was severely pale, mildly icteric, with signs of moderate dehydration. There was no pedal edema, no digital clubbing, nor significant peripheral lymphadenopathy.

Central nervous examination showed a conscious but lethargic boy, pupils were equal and reactive to light both direct and consensually. There were no signs of meningeal irritation, and his muscle tone was normal in all limbs.

Musculo-skeletal examination revealed generalized bone tenderness with a tender swelling over the dorsum of the left foot.

Cardiovascular system examination showed a pulse rate of 118 beats per minute, blood pressure of 110/60mmHg (supine), and a normal first and second heart sound.

Respiratory examination showed a dypsnoeic child, with a respiratory rate of 40 cycles per minute and flaring of the alae nasi with intercostal recessions. Percussion notes were resonant and breath sounds vesicular.

His liver was 8cm palpable, below the right subcostal margin, firm and non-tender. Spleen was 6cm palpable, below the left subcostal margin, firm and non-tender. The kidneys were not ballotable.

Percentage saturation of oxygen using a pulse oximeter was between 88 to 90% during initial evaluation. He was therefore placed on intra-nasal oxygen at 1 litre per minute.

A diagnosis of vaso-occlusive crisis with moderate anaemia in a child with sickle cell anaemia was made. He was admitted into the children emergency unit,and an urgent packed cell volume done, was 19%. He was subsequently transfused with packed red cells. He also received analgesics, and intravenous antibiotics - cephalosporins. Nine hours into admission, on a review, was noted to have developed sudden onset of bilateral orbital swelling with proptosis. He also complained of headache, facial bone pains and was still febrile.

Ophthalmologic examination revealed swollen eyelids, proptosis, conjunctival redness, and pupils which were slowly reactive to light bilaterally. This was the second episode of bilateral orbital swelling and proptosis in the child's life. The first occurrence was two years prior to the present admission, and it resolved without any intervention. With evolving symptoms and signs, a diagnosis of Orbital Compression Syndrome (OCS) was made and intravenous normal saline, was also instituted. The Ophthalmologist and Paediatric Surgeon were invited to urgently review the child. Computed Tomographic Scan of the face and orbit was requested but could not be done due to financial constraints of the parents. Few hours later, the child began bleeding from different orifices with marked difficulty in breathing. He died, despite all resuscitative measures instituted.

III. DISCUSSION

Sickle cell anaemia is a genetic disorder of haemoglobin, in which affected people have distorted red blood cells called haemoglobin S.[1-4] The resultant sickled haemoglobin gets worse, especially on exposure to hypoxia, metabolic acidosis or severe dehydration[1,2]. The sickled red blood cells occludes microcirculation and causes infarction to tissues,leading tointermittent episodes of vaso-occlusive crisis which usually presents as painful episodes in various bones and other systemic complications.

Ophthalmologic complications of sickle cell disease are unusual findings[7] and can affect any part of the eye and surrounding bones with potential loss of vision.[8] There have been some reports of ocular manifestations of sickle cell anaemia in the past decades from several countries of the world.[6,9] Though most reports were observed in children aged 10 to 19 years, some cases had been seen in children as young as 2 years.[9] The presenting male patient was nine years old similar to the most commonly reported age for OCS. The complication was also noted to be commoner in males than females as the index patient was.

Orbital compression syndrome (OCS) is an acute condition characterized by eyelid edema, proptosis, periorbital pain, restriction of extra-ocular motility, with or without decreased visual acuity.[8] It develops from orbital bone infarction which is common in children because of the presence of more marrow space in their orbital bone than is found in adults. Another feature is the formation of orbital or intracranial hematomas.[8]

The presentation is usually sudden, and diagnosis is clinical in any patient presenting with eyelid edema, proptosis, periorbital pain, fever, and ocular or visual disturbances in a sickle cell disease patient with vaso-occlusive crisis.[2,9] Other findings may include hyperemia and eyeball movement limitations.[2,9]Appropriate management requires a thorough evaluation to exclude other hemorrhagic, infectious or neoplastic processes, as well as vigilant ophthalmic monitoring. Supportive care is effective, unless optic nerve dysfunction or large hematomas are present, which would indicate that surgical evacuation is warranted to prevent loss of vision and to speed recovery.[8]

Due to financial constraints, our patient was unable to afford imaging studies that would have identified subperiosteal haematoma formation around the orbit as reported by other authors.[1-6,9-11]In contrast to several reports highlighting good recovery in patients with OCS,[1-6, 9-11] our patient died within a few hours of developing Orbital Compression Syndrome. This rapid progression and financial constraints of the parents made investigations and possible surgical intervention difficult. It is possible that this patient may have had a very large haematoma, which led to rapid worsening of symptoms and eventual death.[12-15]
Expedient diagnosis of orbital compression syndrome in children with sickle cell disease is crucial because this is a potentially sight and life-threatening entity.[3]

IV. CONCLUSION

Orbital compression syndrome from sickle cell anaemia, should always be considered a possible complication in any child that presents with sickle cell anaemia, and managing physicians should be on the look-out for this, especially in resource-poor settings, where many patients lack financial capabilities to afford full panel investigations, orthose, with limited or unavailable hi-tech diagnostic equipments.

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